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Journal

Clinical Orthopaedics and Related Research, 346

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Publication Date

1998

Peer reviewed

Catastrophic Falls in Patients Who Have Fibrodysplasia Ossificans Progressiva

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There have been numerous anecdotal reports of catastrophic falls in patients with fibrodysplasia ossificans progressiva. To determine the incidence of serious morbidity and mortality associated with falls in this patient population, the authors surveyed the 135 patient members of the International Fibrodysplasia Ossificans Progressiva Association and an age and gender matched control group. Eighty-one percent of the fibrodysplasia ossificans progressiva population suffered a fall resulting in injury compared with 44% of the controls. Sixty-seven percent of the falls initiated a painful flareup of fibrodysplasia ossificans progressiva leading to permanent loss of movement in almost all patients. Fifty-four percent of all falls suffered by the fibrodysplasia ossificans progressiva group led to permanent disability compared with 4% of all falls in the control group. Although trauma to the head was a common site of injury in both groups, the in-

jury profile in the fibrodysplasia ossificans progressiva group included traumatic brain injuries, intracranial hemorrhage and death whereas the control group suffered mostly minor soft tissue lacerations. Deficiencies in coordinate gait and protective function likely accounted for the severity of injuries especially to the head in the fibrodysplasia ossificans progressiva population. Precautions are recommended that are intended to minimize the risk of injury without compromising a patient's functional level and independence. These recommendations include limitation of high risk activities, protective head gear, safety improvements in living environments, and augmentation of stabilizing and protective functions.

For patients with fibrodysplasia ossificans progressiva, a rare genetic disorder of heterotopic ossification, falls are common and may lead to permanent disability.^{1,3} To determine the incidence of serious morbidity associated with falls in this patient population, the authors conducted a postal survey of all of the patient members of the International Fibrodysplasia Ossificans Progressiva Association and an age and gender matched control group. This study identifies common sites of injury and mechanisms of falls in the fibrodysplasia ossificans progressiva patient population and recommends precautions in-

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Supported in part by a grant from the International Fibrodysplasia Ossificans Progressiva Association.

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MATERIALS AND METHODS

A detailed survey requesting information on falls was mailed to the 135 patient members of the International Fibrodysplasia Ossificans Progressiva Association. A modified survey omitting questions specific to fibrodysplasia ossificans progressiva was administered to 94 patients and their families who were seen in the outpatient adolescent, dermatology, otolaryngology, general pediatrics, or cardiology clinics one morning at Children's Hospital of Philadelphia and individuals sitting at predetermined tables in a popular local nonhospital cafeteria. Participants in both surveys were asked whether they had ever suffered a fall that resulted in an injury. Specific information about the fall was obtained including the age at time of the fall, the mechanism of the fall (what factors initiated the fall), whether the fall occurred inside the home or outside, and the number of falls that occurred in the year before the survey. Narrative information was obtained about injuries suffered from the fall, and included specific data on whether the patient suffered a head injury, loss of consciousness or blackout, or an injury to the neck, jaw, back, arms, or legs. Each participant provided information on treatment including a narrative description of any emergency care or hospital admission. Patients with fibrodysplasia ossificans progressiva were asked about disease exacerbations and flareups of fibrodysplasia ossificans progressiva caused by the injury. Information was sought on the anatomic areas affected by the flareups, whether movement was lost secondary to the flareups, how much movement was lost, and in what anatomic locations the movement was lost. Outcomes data were obtained on all patients including information on permanent change in joint function or mobility as a result of the fall.

At the time of the survey, the 135 patients represented more than 90% of all known patients in the world who had fibrodysplasia ossificans progressiva. Followup was performed by telephone only in those cases where ambiguous answers existed.

Statistical significance of the difference in likelihood of falling was conducted using the chi square test, and differences in the likelihood of injury were assessed using Fisher's exact test.² All tests were two sided.

RESULTS

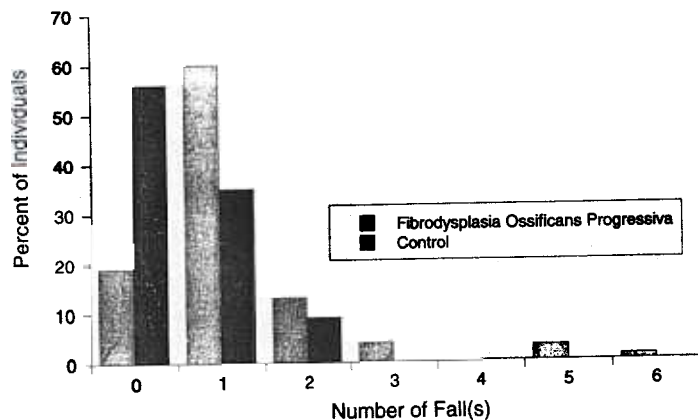
Experimental Group

The postal survey was returned by 112 (83%) of the 135 patients with fibrodysplasia ossificans progressiva. There were 64 (57%) females and 48 (43%) males ranging in age from 2 years to 75 years (median age, 25 years). Ninety-one (81%) of the 112 patients had suffered at least one fall with resulting injury. Twenty-one (19%) patients had not suffered any falls (Fig 1). One hundred thirty-three falls resulting in injuries were reported by the 91 patients. The median age at the time of a fall with injury was 12 years (range, 0.1-75 years). Sixty percent of the falls with injury occurred outside of the home, whereas 40% occurred inside the home (Table 1).

The mechanisms that initiated the falls were grouped into nine general categories (Fig 2). The most common mechanism causing a fall was loss of balance or tripping (31%), followed by playground or sports mishap (16%), loss of traction on ice or other surfaces (13%), uneven flooring (11%), a bicycle or motorcycle mishap (9%), a misstep on stairs (8%), imbalance during transfer from chair or bed (7%), collisions with others (4%), and falling from a wheelchair (1%).

Injury to the head was the most common anatomic site of injury and occurred in 57 falls (43%), (Table 2). Listed among the 26 (20%) severe head injuries were loss of consciousness in 23 (17%) falls concussion in 13 (10%) falls, intracranial hemorrhage in five (4%) falls, depressed skull fracture in three (2%) falls, and focal neurologic deficits in three (2%) falls. Minor head injuries included soft tissue lacerations in (11%) falls and contusions in 23 (17%) falls. Injuries to the neck occurred in 34 (26%) falls, and fractures of either heterotopic bone in the neck or cervical vertebrae occurred in nine (7%) falls. Injuries to the jaw occurred in 15 (11%) falls, back or chest wall in 41 (31%) falls, upper limbs in 53 (40%) falls, and lower limbs in 54 (41%). Fracture of either normotopic or heterotopic bone occurred in 30 (23%) of the falls.

Fig 1. Frequency of falls. Ninety-one (81%) of the 112 patients with fibrodysplasia ossificans progressiva suffered at least one fall with resulting injury, whereas only 41 (44%) of the 93 control patients suffered at least one fall with resulting injury.



The outcomes of all falls are listed in Table 3. Ninety-nine percent of the patients survived their falls. Five patients in this study suffered intracranial hemorrhage requiring neurosurgical intervention. One patient died secondary to severe head trauma suffered when she fell while attempting to dance. One patient remained in a coma for 2 weeks after a fall, two patients had a seizure disorder develop. In 68% of the falls, the patients were seen in the emergency department. Twenty-one percent of the falls resulted in a hospital admission. Flareups of fibrodysplasia ossificans progressiva were induced by 67% of falls. Ninety-three percent of these flareups caused loss of movement. Fifty-four percent of the falls resulted in a permanent change of function. In falls

that caused a flareup of fibrodysplasia ossificans progressiva, 79% resulted in a permanent change of function. A permanent change in the ability to walk occurred after 38% of all falls. However, 78% of the falls in which the lower limb was affected by a flareup were followed by a permanent change in the ability to walk.

Control Group

In the control group, 93 (95%) of the 98 people asked to participate in the study completed a survey form. There were 51 (55%) females and 42 (45%) males ranging in age from 2 years to 75 years (median age, 28 years). Forty-one (44%) of the 93 participants had suffered at least one fall resulting in an injury. Forty-nine falls resulting in in-

TABLE 1. Demographic Profiles

Parameter	Fibrodysplasia Ossificans Progressiva	Control
Participating individuals	112 (83%)	93 (95%)
Median age (years) (range)	25 (2-75)	28 (2-75)
Gender ratio (females/males)	1.3:1	1.2:1
Median age (years) at time of fall that resulted in an injury (range)	12 (0.1-75)	13 (2-68)
Individuals suffering at least one fall that resulted in an injury	91 (81%)	41 (44%)
Falls that resulted in injury	133	49
Falls inside home	80 (60%)	31 (63%)
Falls outside home	53 (40%)	18 (37%)

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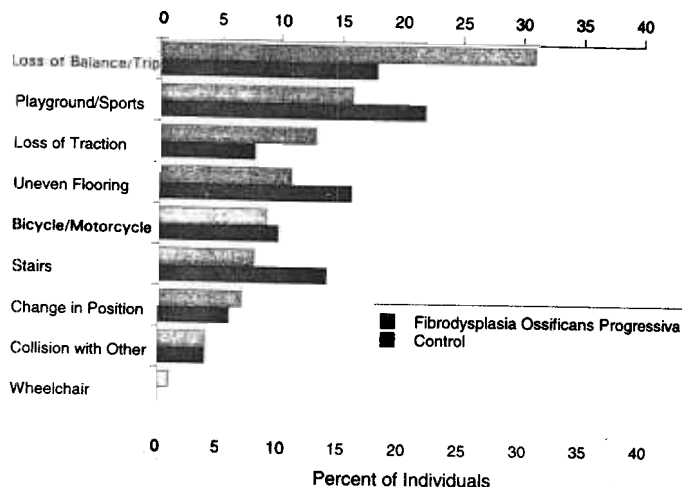


Fig 2. Mechanisms of falls. The mechanisms that initiated the falls were grouped into nine general categories. In the fibrodysplasia ossificans progressiva, the most common mechanism causing a fall was loss of balance or tripping (31%), followed by a playground or sports mishap (16%), loss of traction on ice or other surfaces (13%), uneven flooring (11%), a bicycle or motorcycle mishap (9%), a misstep on stairs (8%), imbalance during transfer from chair or bed (7%), collisions with others (4%), and falling from wheelchair (1%). In the control group, the most common mechanism causing a fall was playing at a playground or engaging in a casual sporting activities (22%). Loss of balance (18%), uneven flooring (16%), a misstep on stairs (14%), fall from a bicycle or motorcycle (10%), loss of traction on ice or other surfaces (8%), change in position including fall from bed (6%), and collisions with others (4%) were responsible for more than 98% of all falls.

juries were reported by the 41 people (Fig 1). The median age when a fall occurred was 13 years (range, 2–68 years). Sixty-three percent of the falls with injury occurred outside of the home; 37% occurred inside the home (Table 1).

The most common mechanism causing a fall was playing at a playground or engaging in casual sporting activities (22%), (Fig 2). Loss of balance (18%), uneven flooring (16%), a misstep on stairs (14%), a bicycle or motorcycle mishap (10%), loss of traction on ice or other surfaces (8%), change in position including fall from bed (6%), and collisions with others (4%) were responsible for more than 98% of all falls.

Head involvement was mentioned in 16 (33%) falls with patients suffering loss of consciousness in two (4%) falls, minor soft tissue lacerations in 15 (31%) falls, or contusions in four (8%) falls. Neck injury oc-

curred in two (4%) falls. Injuries to the jaw were suffered in two (4%) falls, back or chest walls in four (8%) falls, and limbs in 30 (61%) falls. A fracture occurred in 17 (35%) falls (Table 2).

In 61% of the falls, the patients were seen in the emergency department. Eight percent of the falls resulted in a hospital admission. Four percent of all of the falls resulted in a permanent change of function. A permanent change in the ability to walk occurred after 2% of all falls (Table 3).

Comparisons

In the fibrodysplasia ossificans progressiva group, the percentage of respondents who reported a fall that resulted in an injury (81%) was much larger than the percentage of respondents who reported a fall in the control group (44%), and was highly significant ($p = 7 \times 10^{-8}$). The most significant difference be-

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TABLE 2. Injury Profile From Falls

Injury	Injury Rate as a Result of Falls	
	Fibrodysplasia Ossificans Progressiva (%)	Control (%)
Head	43	32
Severe head injuries	20	4*
Forehead/head laceration	11	31*
Neck	26	4*
Fractures of neck	7	0
Soft tissue injury	4	4
Jaw injury	11	4
Back/torso injury	31	8*
Fractures	1	4
Soft tissue injury	9	4
Upper extremity	40	35
Fractures	13	20
Heterotopic ossification	1	0
Shoulder/humerus	6	4
Elbow/forearm	5	12
Wrist/finger	2	4
Soft tissue injury	10	8
Lower extremity	41	27
Fractures	9	10
Hip/femur	4	0
Knee/tibia	1	0
Ankle/foot	4	10
Soft tissue injury	14	20

*Significant difference. $P < 0.05$.

tween the fibrodysplasia ossificans progressiva group and the control group was a greater frequency in patients with fibrodysplasia ossificans progressiva of severe head injury ($p = .01$), loss of consciousness ($p = .017$), concussion ($p = .021$), neck injury ($p = .001$), and back or torso injury ($p = .002$). The control group had a significantly greater frequency of forehead or head lacerations ($p = .003$). There was no significant difference in the overall frequency of head trauma ($p = .326$).

In disposition and outcomes, the group of patients with fibrodysplasia ossificans progressiva was more likely to be admitted to a hospital ($p = .048$), to have had a permanent change of function because of a fall ($p = 0$), and to have had a permanent change in the ability to walk because of a fall ($p = 0$). The difference in the rate of emergency room admissions was not significant.

DISCUSSION

The results of this study clearly establish the catastrophic nature of falls suffered by the fibrodysplasia ossificans progressiva population compared with an age and gender matched control group. Patients with fibrodysplasia ossificans progressiva suffered an impressively greater number of falls resulting in injury than the people in control group suffered: 81% in the fibrodysplasia ossificans progressiva group compared with 44% in the control group. The injury profile among the fibrodysplasia ossificans progressiva group was also considerably more severe. As disturbing, 2/3 of the falls initiated a painful flareup of disease activity leading to permanent loss of movement. Overall, more than half of all falls in the fibrodysplasia ossificans progressiva population led to permanent disability.

TABLE 3. Disposition and Outcomes of Individuals Suffering a Fall

Disposition and Outcomes	Fibrodysplasia Ossificans Progressive (%)	Control (%)
Emergency room visit	68	61
Hospital admission	21	8
Flare induced by fall	67	NA
Loss of movement caused by flareup	93	NA
Permanent change in function resulting from fall	54	4
Permanent change in function if a flareup occurred secondary to fall	79	NA
Permanent change in ability to walk resulting from fall	38	2
Permanent change in ability to walk of a flareup induced by fall affected lower extremity including hips	78	NA

NA = not applicable.

There was no significant difference in the number of head injuries suffered by the two groups. However, catastrophic head injuries were clearly more common in patients with fibrodysplasia ossificans progressiva, whereas minor lacerations of the scalp were more common in the control group. Protective functions often limit injury when a fall occurs in a neurologically and biomechanically unimpaired individual. However, patients with fibrodysplasia ossificans progressiva do not have normal mechanisms to protect themselves during a fall. As a result, patients suffer a disproportionate number of severe injuries to the head because of early loss of upper limb protective function at a time when patients still remain precariously ambulatory. Many patients frequently are trapped in a cycle of falls, injuries, disabilities, and subsequent falls (Fig 3).

Measures to prevent falls and associated life threatening injuries in patients with fibrodysplasia ossificans progressiva should be directed at modification of activity, improvement in household safety, augmentation of gait, and use of protective headgear. Redirection of activity to less interactive play may provide an alternative solution. Complete avoidance of high risk circumstances may re-

duce falls, but also may compromise a patient's functional level and independence, and may be unacceptable to some. Within the home, adjustments to the living environment to reduce the number of falls may include installing protective hand railings on stairs, securing loose carpeting, removing objects from walkways, and eliminating uneven flooring

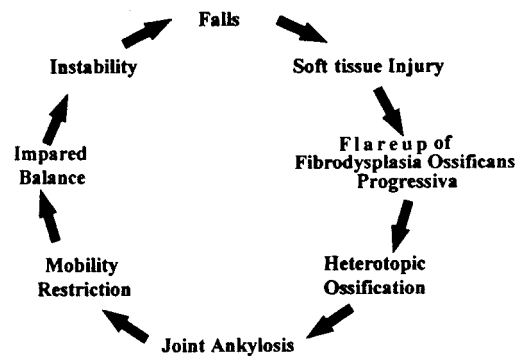


Fig 3. Self-perpetuating fall cycle in patients who have fibrodysplasia ossificans progressiva. Minor soft tissue trauma can lead to severe exacerbations of fibrodysplasia ossificans progressiva with resultant heterotopic ossification and joint ankylosis. Mobility restriction from joint ankylosis severely impairs balancing mechanisms, causing instability, resulting in subsequent falls.

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including thresholds of door frames. Prevention of falls due to imbalance begins with stabilization of gait. The use of a cane or stabilizing device may improve balance for many patients. For more mobile individuals, the use of a rolling cane or walker will assist in stabilization.

Augmentation of the patient's protective functions should be performed to minimize injury when a fall does occur. Implementation of helmet use in young patients with fibrodysplasia ossificans progressiva may help reduce the disturbing incidence of severe head injuries.

When a fall occurs, prompt medical attention should be sought, especially when a head injury is suspected. Any head injury should be considered serious until proven otherwise. A few common signs and symptoms of severe head injury include increasing headache, dizziness, drowsiness, obtundation, weakness, confusion, or loss of consciousness. These symptoms often do not appear until hours after an injury. A patient should be examined carefully by a healthcare professional if any head injury is even remotely suspected.

Several biases may have been introduced in this study. First, the authors relied on the memories of patients and their families, and on volunteer controls to generate the data for this study, therefore introducing recall bias. Although the total recall of all falls may be in question, the recall of those falls associated with severe injury and lifelong disability are likely to be remembered with great accuracy and fidelity in the fibrodysplasia ossificans progressiva group and in the age matched and gender matched control group. Second, despite the extraordinary response

of more than 80% in the fibrodysplasia ossificans progressiva patient population, some questionnaires were not returned. It is unlikely, however, that these questionnaires would have altered the final conclusions significantly. Third, the control group was matched for age and gender only, and no attempt was made to take into account activity level, socioeconomic status, or detailed medical history. Injuries that were suffered while participating in an organized athletic activity also were excluded. Overall, the fibrodysplasia ossificans progressiva group leads a more sedentary lifestyle than the age and gender matched control group.

This study clearly establishes the catastrophic nature of falls in patients afflicted with fibrodysplasia ossificans progressiva. Through activity modification, improved household safety, and gait stabilization, the number of falls suffered in this group could be reduced. However, despite aggressive preventive measures, falls will occur. When a fall occurs, augmentation of a patient's protective functions such as use of protective head gear may minimize injury. Early recognition of severe injuries, especially those related to head trauma, may lead to a reduction in morbidity and mortality.

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